

MONITOR

A Semi-Annual Data and Research Update
Texas Department of Health, Bureau of Epidemiology



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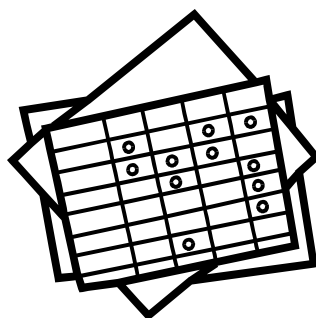
FROM THE DIRECTOR

THE TEXAS BIRTH DEFECTS MONITORING DIVISION APPROACHES MILESTONE

On March 21, 1994, the Texas Birth Defects Monitoring Division began operations. As the first employee of this program, the approaching 10-year anniversary provokes many reminiscences and much pride and gratitude in the work that has been done, and to the many professionals who have contributed to our successes over the years. This milestone also brings to mind the enormous amount of work before us in our quest to better understand the causes of birth defects and to affect their prevention.

Some significant milestones include:

- April 1991 Anencephaly cluster identified in Brownsville
- June 1993 Birth Defects Act signed into law
- September 1993 Birth Defects Act went into effect
- March 1994 Divisional offices opened in Austin
- December 1994 Data collection begun in Texas Birth Defects Registry pilot regions
- April 1995 First issue of *Texas Birth Defects Monitor* published
- October 1996 Texas Birth Defects Research Center funded and established



- January 1997 Registry expanded from 35% to 80% of all live births
- April 1997 Scientific and community forum on neural tube defects held in Brownsville
- September 1997 Studies initiated for the Texas Birth Defects Research Center
- October 1997 Published first annual report based on Registry data
- January 1998 Registry expanded statewide
- November 1999 Texas receives its "A" rating for birth defects registries from the Pew Commission
- November 2002 Statewide data first published
- November 2003 Referral component implemented

One of our proudest accomplishments has been nine straight years of this publication, now the *Texas Birth Defects Monitor: A Semi-Annual Data and Research Update*. Because many of the issues contain special data reports and other features that serve as reference material, we are now providing an index to topics, which is available on our web site at www.tdh.state.tx.us/tbdmd/monitor/the.htm. Requests for this index, back issues, or suggestions for future topics can be sent to Amy Case, Editor, at 512-458-7232 or amy.case@tdh.state.tx.us.

WHAT'S INSIDE

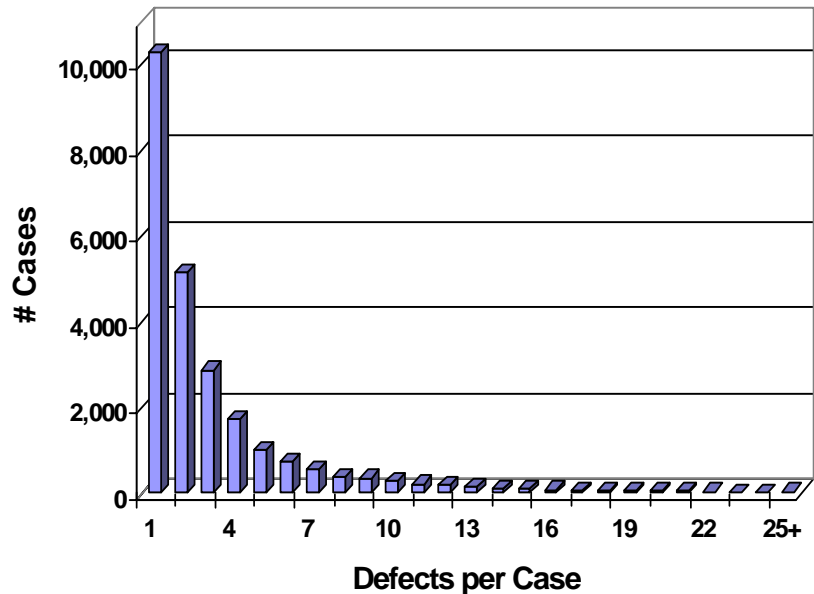
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FROM THE REGISTRY

MAJORITY OF INFANTS AND FETUSES IN THE REGISTRY HAVE MORE THAN ONE DEFECT

BIRTH DEFECTS PER CASE IN THE TEXAS BIRTH DEFECTS REGISTRY: Rates reported from the Texas Birth Defects Registry reflect the number of occurrences of each defect or defect category. However, the number of birth defects cannot be summed to get the total number of children born with birth defects in Texas each year, for the simple reason that many of these infants or fetuses will have more than one defect (See Chart). About 42% of all deliveries in the Registry have just one defect (isolated defect), while 21% had two birth defect diagnoses. Thirty-seven cases had 25 or more monitored diagnoses.

Distribution of number of birth defect diagnoses per case, Texas, Birth Defects Registry, 1999 and 2000

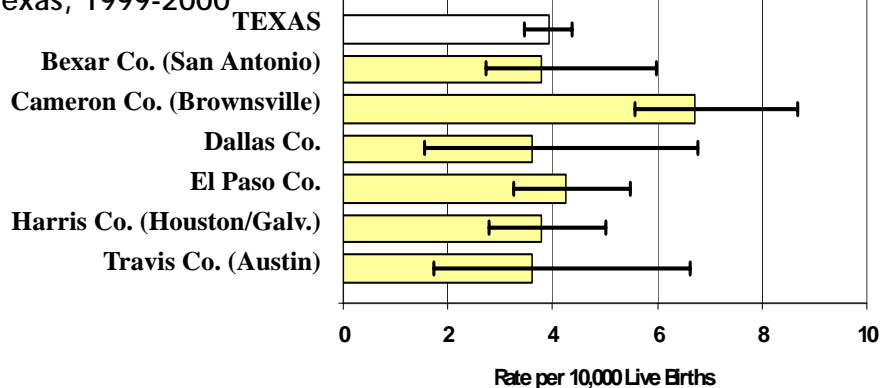


Note: Does not include possible/probable birth defects diagnoses.

SELECTED DEFECTS BY COUNTY

With two combined years of birth defects data for every county in Texas, increasingly reliable comparisons can be made between various counties and regions, as in the charts below. Overlapping error bars on the charts below indicate that the differences between two rates do not meet statistical significance. These comparisons should be viewed with caution, because all prevalence rates are crude estimates (unadjusted for maternal characteristics such as race/ethnicity or age).

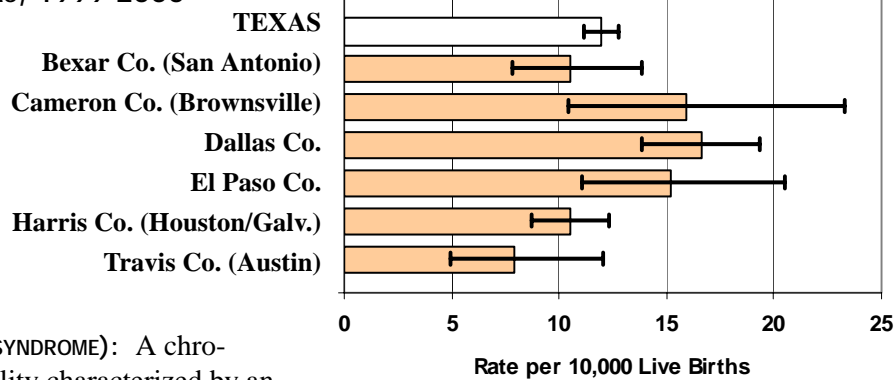
Gastroschisis, Texas, 1999-2000



GASTROSCHISIS: A congenital opening of the abdominal wall with protrusion of the intestines. This condition is surgically treated.

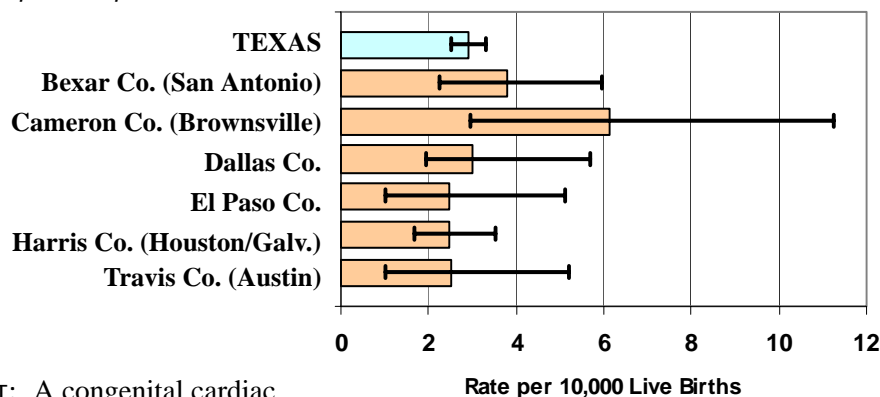
For more information on birth defect rates by county, contact the Texas Birth Defects Monitoring Division at 512-458-7232 or birthdefects@tdh.state.tx.us.

Trisomy 21, Texas, 1999-2000



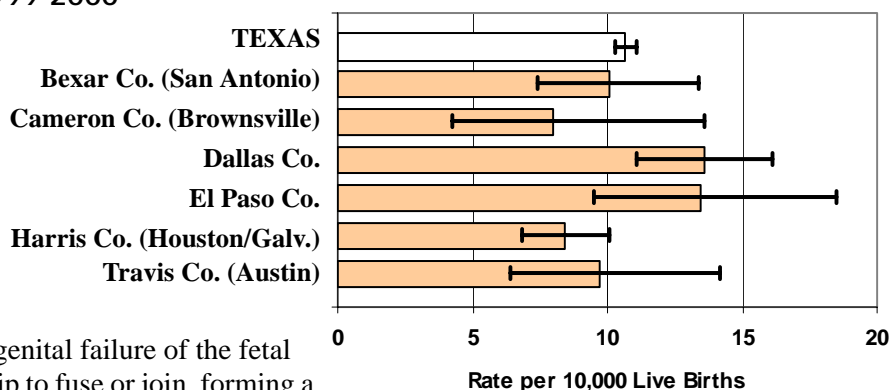
TRISOMY 21 (DOWN SYNDROME): A chromosomal abnormality characterized by an extra copy of chromosome 21. In rare cases this syndrome is caused by translocation. Down syndrome is characterized by moderate to severe mental retardation, sloping forehead, small ear canals, flat-bridged nose and short fingers and toes. One third of infants have congenital heart disease, and one third have duodenal atresia. (Both can be present in the same infant.) Affected people can survive to middle or old age.

Tetralogy of Fallot, Texas, 1999-2000



TETRALOGY OF FALLOT: A congenital cardiac anomaly consisting of four defects: ventricular septal defect, pulmonary valve stenosis or atresia, displacement of the aorta to the right, and hypertrophy of right ventricle. The condition is corrected surgically.

Cleft Lip with or without Cleft Palate, Texas, 1999-2000



CLEFT LIP: The congenital failure of the fetal components of the lip to fuse or join, forming a groove or fissure in the lip. Infants with this condition can have difficulty feeding, and may use assistive devices for feeding. This condition is corrected when the infant can tolerate surgery. **Cleft palate:** The congenital failure of the palate to fuse properly, forming a grooved depression or fissure in the roof of the mouth. This defect varies in degree of severity. The fissure can extend into the hard and soft palate and into the nasal cavities. Infants with this condition have difficulty feeding, and may use assistive devices for feeding. Surgical correction is begun as soon as possible.

DIFFERENCES IN DEFECT RATES BETWEEN MALE AND FEMALE INFANTS/FETUSES

Certain defects show markedly different patterns between male and female offspring. While the reason for the differences is obvious in some cases (as with hypospadias among males, or X-linked autosomal syndromes for females), others are not so obvious and may help to generate hypotheses about the underlying causes of these defects. Patterns we have observed in Texas are presented below. Of particular interest is that for all monitored malformations, males are approximately 40% more likely to be affected (3.9% of all deliveries vs. 2.8% for females.) Other studies have found similar differences in birth defect rates*.

Prevalence of Selected Birth Defects by Sex of Infant or Fetus, Texas, 1999-2000

Birth Defects more Prevalent among Males than Females

Defect	Sex [†]	Cases	Rate [†]	95% Confidence Interval for Rate	
Hydrocephaly (p=0.0126)	Male	307	8.43	7.49	- 9.38
	Female	237	6.80	5.94	- 7.67
Transposition of the great vessels (p<0.0001)	Male	209	5.74	4.96	- 6.52
	Female	128	3.67	3.04	- 4.31
Aortic valve stenosis (p=0.0012)	Male	101	2.77	2.23	- 3.32
	Female	57	1.64	1.24	- 2.12
Hypoplastic left heart syndrome (p=0.0413)	Male	83	2.28	1.82	- 2.83
	Female	56	1.61	1.21	- 2.09
Cleft lip with or without cleft palate (p<0.0001)	Male	463	12.72	11.56	- 13.88
	Female	295	8.47	7.50	- 9.43
Pyloric stenosis (p<0.0001)	Male	1103	30.30	28.51	- 32.09
	Female	257	7.38	6.47	- 8.28
Hirschsprung disease (p=0.0003)	Male	60	1.65	1.26	- 2.12
	Female	25	0.72	0.46	- 1.06
Hypospadias or epispadias (p<0.0001)	Male	2042	56.09	53.66	- 58.53
	Female	1	0.03	0.00	- 0.16
Renal agenesis or dysgenesis (p=0.0002)	Male	215	5.91	5.12	- 6.70
	Female	137	3.93	3.27	- 4.59
Obstructive genitourinary defect (p<0.0001)	Male	976	26.81	25.13	- 28.49
	Female	410	11.77	10.63	- 12.91
Craniosynostosis (p<0.0001)	Male	174	4.78	4.07	- 5.49
	Female	98	2.81	2.28	- 3.43
Infants and fetuses with any monitored birth defect (p<0.0001)	Male	14247	391.36	384.93	- 397.78
	Female	9820	281.83	276.25	- 287.40

Birth Defects more Prevalent among Females than Males

Trisomy 21 (Down syndrome) (p=0.0103)	Male	469	12.88	11.72	- 14.05
	Female	376	10.79	9.70	- 11.88
Ventricular septal defect (p<0.0001)	Male	1353	37.17	35.19	- 39.15
	Female	1533	44.00	41.79	- 46.20
Cleft palate alone (without cleft lip) (p=0.0102)	Male	195	5.36	4.60	- 6.11
	Female	239	6.86	5.99	- 7.73
Biliary atresia (p=0.0105)	Male	17	0.47	0.27	- 0.75
	Female	34	0.98	0.68	- 1.36
Congenital hip dislocation (p<0.0001)	Male	100	2.75	2.24	- 3.34
	Female	262	7.52	6.61	- 8.43

[‡]excludes infants and fetuses with undetermined sex

[†]cases per 10,000 live birth

*Lary JM, Paulozzi LJ. Sex differences in the prevalence of human birth defects: a population-based study. Teratology. 2001 Nov;64(5):237-51.

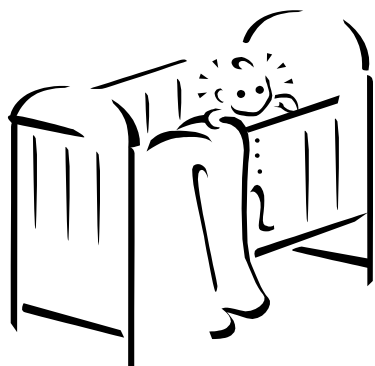
*James WH. Comment on: Teratology. 2001 Nov;64(5):237-51. Potential of the hormonal hypothesis of sex ratio to explain the unusual sex ratios typical of some malformations. Teratology. 2002 Sep;66(3):101-2; author reply 103-4.

PREVENTION

INADEQUATE PREGNANCY WEIGHT GAIN AND BIRTH DEFECTS

Some studies have shown an association between low maternal weight gain during pregnancy and certain birth defects, including neural tube defects, renal agenesis, oral clefts, and microcephaly. It should be noted that the co-existence of low pregnancy weight gain and birth defects does not necessarily indicate that the one causes the other. However, adequate weight gain (usually considered between 15-40 pounds, depending on mother's pre-pregnant weight) in pregnancy is one reliable indicator of maternal health and good pregnancy outcomes.

In the following Texas counties, in greater than 12% of live births the mother gained less than 15 pounds during her pregnancy. (The rate for all of Texas was 10.2%.)



Texas Counties with Average Pregnancy Weight Gain <15 lbs., 1997-2001

County	Number of Births*	% of All Live Births*
Val Verde	1,215	48.4
Kinney	51	40.8
Coryell	1,025	22.3
Bell	5,039	21.2
Willacy	265	19.7
Sherman	33	19.5
Cameron	6,017	19.3
Edwards	17	18.5
Brooks	112	17.7
Dickens	20	17.5
McLennan	2,563	16.8
Sutton	44	16.7
Rusk	427	15.8
Nacogdoches	598	15.3
Hudspeth	22	15.1
Briscoe	17	15.0
Falls	132	14.9
Moore	234	14.9
Zavala	147	14.8
Shelby	248	14.7
Colorado	177	14.5
Jim Hogg	56	14.5
Dimmit	121	14.3
Foard	10	14.1
Howard	295	14.1
Presidio	85	14.0
Victoria	899	14.0
Zapata	152	14.0
San Augustine	72	13.9
Haskell	44	13.8
Kent	4	13.8
Limestone	184	13.8
Hall	36	13.7
Webb	3,177	13.4
Reagan	31	13.3
Reeves	101	13.3
Hill	266	13.2
Uvalde	287	13.0
Delta	36	12.9
Pecos	137	12.9
Gonzales	163	12.8
Hidalgo	6,884	12.8
Hunt	585	12.8
Live Oak	73	12.8
Swisher	83	12.8
Jackson	119	12.7

RESEARCH CENTER

RESEARCH SYMPOSIUM BRINGS DATA BACK TO THE COMMUNITY

In 1996, a Texas Birth Defects Symposium was held in Brownsville with a primary focus on the tracking and reporting of high neural tube defect (NTD) rates along the Texas-Mexico border. In the eight years since, the Texas Center for Birth Defects Research and Prevention (TCBD RP) was established, the Texas Birth Defects Registry became statewide, and active research collaborations have developed to gain understanding into birth defect causes and prevention.

On April 8, 2004, a Birth Defects Symposium will be held in San Antonio (Omni Hotel) to bring the data and research full circle. Presentations will include the most recent findings from the Texas Neural Tube Defects Project, a comparison of selected birth defect rates among Texas counties and regions, and a look at risk factors associated with oral clefts in Texas. A scientific poster session spotlighting student work in birth defects epidemiology and research will also be featured.

Those encouraged to attend this free event include state and local public health professionals, clinicians, researchers from other fields, and anyone with a personal or professional interest in birth defects. For more information about this event, contact Amy Case at 512-458-7232, amy.case@tdh.state.tx.us. Reservations for the Omni Hotel can be made by following the link at our web page, www.tdh.state.tx.us/tbdmd/index.htm, or by calling 210-691-8888 (specify Texas Birth Defects Monitoring Meeting; all rooms are \$80/night plus taxes).

RECENTLY PUBLISHED MANUSCRIPTS BY COLLABORATORS AND COLLEAGUES

Felkner M et al. *Diarrhea: A new risk factor for neural tube defects?* Birth Def Res Part A. 2003;67:504-8.

Suarez L, Cardarelli K, Hendricks, K. *Maternal stress, social support, and risk of neural tube defects among Mexican Americans.* Epidemiology 2003 Sep;14(5):612-6.

Volcik KA, Shaw GM, Lammer EJ, Zhu H, Finnell RH. *Evaluation of infant methylenetetrahydrofolate reductase genotype, maternal vitamin use, and risk of high versus low level spina bifida defects.* Birth Defects Res Part A Clin Mol Teratol.; 67(3): 154-7.

HELLOS AND GOOD-BYES

The Texas Center for Birth Defects Research and Prevention welcomes our new Research Specialist, Tunu Ramadhani, Ph.D. Dr. Ramadhani conducts statistical analyses for the National Birth Defects Prevention Study, and local studies. Her special areas of interest include research methodology, statistical analyses, economics of birth defects, and socio-cultural aspects of birth defects. Dr. Ramadhani can be reached at 512-458-7232, tunu.ramadhani@tdh.state.tx.us.

We regret to announce the Dr. Dawna Wright, who has been the Grant Coordinator for the TCBDRP since its inception in 1996, has departed for a position in the pharmaceutical industry. Thanks and best wishes go out to Dr. Wright.

Inquiries about the open positions at the TBDMD Central Office may be addressed to Mark Canfield, Ph.D. at mark.canfield@tdh.state.tx.us.

NEWBORN HEARING SCREENING UPDATE

SOMETIMES PILOT PROJECTS WORK

AN UPDATE ON THE "SOUNDS OF TEXAS": In 1997, the Audiology Services Program in the Bureau of Children's Health at TDH reported on a pilot project called the "Sounds of Texas," a public-private partnership to implement newborn hearing screening (NBHS) in Texas hospitals. From 1996 to 1999, 80,000 newborns were screened for hearing loss at 30 Texas hospitals, and the "Sounds of Texas" became a national model. The three-year pilot also provided impetus for the passage of House Bill 714, mandating NBHS programs at Texas birth facilities and designating TDH as the oversight agency.

Fast forward to 2003 – the State's universal NBHS program, now known as the Texas Early Hearing Detection and Intervention (TEHDI) Program, oversees NBHS program implementation at 196 birth facilities. Some 300,000 babies are screened for hearing loss at these facilities each year.

The driving force behind the initial pilot project and the resulting mandated program was the need to screen babies for hearing loss at an early age. Historically, in Texas, children with hearing impairment and deafness were not being identified until they were around 56 months old. Recognizing that hearing loss in a baby's first six months of life has been shown to be critical to the development of his language and learning abilities, TDH knew they had to take this window of opportu-

nity to help children with hearing loss achieve optimal academic, communication and social functioning. While early hearing screening was the first step, TDH quickly learned that it needed to go further. Once the baby was screened, there had to be a system in place for further testing if needed and referral to community services. With this in mind, TDH developed a screening and intervention process, adhering to Health and Safety Code, Chapter 47 and recommendations from the American Academy of Pediatrics Position Statement, the Joint Committee on Infant Hearing 2000 Position Statement, and the TDH Audiologic Evaluation Protocol. The "Newborn Hearing 1-3-6 Month Process Practitioner's Guide," available at www.tdh.state.tx.us/audio/newborn-hear.htm, outlines all the steps taken to assure Texas newborns are properly screened for hearing loss, re-tested as needed, and referred for evaluation, intervention and education. This important tool, along with program rules and NBHS program certification criteria developed by TEHDI, serve as quality assurance markers.

To help its 196 NBHS programs be successful, TEHDI provides birth facilities with training, information management and tracking software, and technical assistance as needed. In fact, TDH will soon migrate to a new state-of-the-art web based system that will provide a direct link from the facility's hearing screen equipment to provide accurate and immediate results to the State.

Texas continues to receive recognition for its NBHS program. For the second consecutive year, TEHDI received a rating of Excellent by the National Campaign for Hearing Health (NCHH). The NCHH, in partnership with the American Acad-

emy of Pediatrics and the National Center for Hearing Assessment and Management, issues an annual report card rating state NBHS program. The Excellent rating means that almost all babies in Texas are screened for hearing loss during the birth admission, and a statewide system for coordination, training, quality assurance, and follow-up is in place. Texas screened more than 97% of babies born last year, compared to the national average of 86%. About 5% of babies screened for hearing loss are referred for further evaluation. Of those babies referred, 3 out of 1,000 are typically diagnosed with permanent hearing loss.

For more information about TEHDI, please contact Elaine Braslow, program coordinator, at 512-458-7111, extension 3829 or elaine.braslow@tdh.state.tx.us.

| ANNOUNCEMENTS

STAFF CHANGES AND MORE AT TBDMD

As of August 31, 2003, Staff Services Officer Sandy Wicker retired. Sandy was one of the first three employees on staff at the Texas Birth Defects Monitoring Division, providing administrative support as well as her constant commitment and hard work for nine years. Many thanks to Sandy!

In addition, the new fiscal year ushered in many changes in our organizational chart. As of September 1, 2003, supervision of the regional surveillance staff moved to Central office oversight. Dan Driggers, formerly Quality Control manager, is now Regional Operations Manager. Region 8 (San Antonio/South Texas are) and Region 11 (Lower Rio Grande Valley) have been merged under the supervision of Jorge Trevino, who has ably conducted Region 11 birth defects surveillance activi-

ties since the inception of the first pilot phase. Finally, Ann Phelps, who has also been with TBDMD since the beginning, has moved from Region 8 to Central Office, where she is coordinating quality control activities with staff training efforts.

We are already seeing benefits of these changes in streamlined processes, improved data collection procedures, and exciting new plans for improving staff training.

MARCH OF DIMES LICENSE PLATES AVAILABLE

On June 22, 2003, Governor Perry signed House Bill 2971 into law. Among other things, the legislation allowed for the production of March of Dimes specialty license plates. The license plates will cost an additional \$30 above a normal renewal or purchase price, of which \$8 will go towards administrative costs and \$22 will support the Texas Birth Defects Registry. Support of the Texas Birth Defects Registry is a March of Dimes public affairs priority. The March of Dimes license plates will go on sale in January 2004. The design will be posted on the Texas Department of Transportation's web site at www.txdot.state.tx.us/vtr/spplates/allplates.htm.

CONTRACEPTIVE WITH FOLIC ACID MAY REDUCE BIRTH DEFECTS

A U.S. advisory panel has determined that adding folic acid to oral contraceptives may be a worthwhile way to further reduce certain types of birth defects, Johnson & Johnson, a leading contraceptive maker, wants to develop prescription-only pills that combine the two. The goal is to reduce the risks of birth defects in babies of women who become pregnant while taking birth control pills, as well as others who conceive shortly after stopping pill use.

A combination pill would be "a convenient and effective way... to reduce the number of neural tube defects in this country with negligible, if any, safety concerns," Dr. Andrew Friedman, director of women's health care research at Johnson & Johnson, told the Food and Drug Administration advisory panel.

An estimated 1 million U.S. women taking birth control pills become pregnant each year because they do not take the contraceptives properly. And many of them do not consume the recommended amount of folic acid to prevent neural tube defects, in which the tube that later becomes the brain and central nervous system does not close properly, Friedman said. Research has indicated that plasma folate levels tend to drop gradually over the course of three months after discontinuation of an oral contraceptive/folic acid product. Therefore, a woman would have better folate status should she conceive shortly after discontinuing the pills than if she had not been taking any folic acid supplement.

Since 1996, U.S. health officials recommend women capable of becoming pregnant consume 400 micrograms of folic acid per day through supplements or diet to prevent neural tube defects. Because the neural tube closes about four weeks after conception -- before many women know they are pregnant -- folic acid needs to be taken before a woman conceives to be beneficial.

Members unanimously endorsed the concept of combining birth control pills with folic acid, but they said more research was needed to address various concerns.

The Johnson and Johnson briefing to the FDA can be found at www.fda.gov/ohrms/dockets/ac/03/briefing/4002B1_01_Johnson-Johnson-Background.pdf.

CALENDAR

2004

- ◆ April 8: Texas Birth Defects Research Symposium, San Antonio. Contact Amy Case, 512-458-7232, amy.case@tdh.state.tx.us. More information at www.tdh.state.tx.us/tbdmd/index.htm.
- ◆ April 14, 3:00 PM - 5:00 PM ET Satellite Broadcast: Reproductive Health Issues for Women Over 40. Contact (334) 206-5618 or visit www.adph.org/alphtn.
- ◆ April 15-18: Texas Genetics Society 31st Annual Meeting, South Padre Island. Contact: 979-862-4775, email jderr@cvm.tamu.edu
- ◆ June 3-6: Texas Health Information Management Association Annual Convention, Houston www.txhima.org/calupcoming.htm

The *Monitor* is published twice a year by the Texas Birth Defects Monitoring Division, Texas Department of Health.

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To be added to our mailing list and for other free publications from the Division, please contact us at 512-458-7232 or e-mail bobbie.mankowski@tdh.state.tx.us.

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